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Cavopulmonary assist for the failing Fontan circulation: Impact of ventricular function on mechanical support strategy

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Abstract

Mechanical circulatory support - either ventricular assist device (VAD, left-sided systemic support) or cavopulmonary assist device (CPAD, right-sided support) - has been suggested as treatment for Fontan failure. The selection of left- vs. right-sided support for failing Fontan has not been previously defined.

Computer simulation and mock circulation models of pediatric Fontan patients (15–25 kg) with diastolic, systolic, and combined systolic and diastolic dysfunction were developed. The global circulatory response to assisted Fontan flow using VAD (HeartWare HVAD, FL) support, CPAD (Viscous Impeller Pump, IN) support, and combined VAD and CPAD support were evaluated.

Cavopulmonary assist improves failing Fontan circulation during diastolic dysfunction but preserved systolic function. In the presence of systolic dysfunction and elevated ventricular end-diastolic pressure (VEDP), VAD support augments cardiac output and diminishes VEDP, while increased preload with cavopulmonary assist may worsen circulatory status.

Fontan circulation can be stabilized to biventricular values with modest cavopulmonary assist during diastolic dysfunction. Systemic VAD support may be preferable to maintain systemic output during systolic dysfunction. Both systemic and cavopulmonary support may provide best outcome during combined systolic and diastolic dysfunction. These findings may be useful to guide clinical cavopulmonary assist strategies in failing Fontan circulations.

Keywords

Failing Fontan; cavopulmonary support; VAD support; systolic dysfunction; diastolic dysfunction

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Introduction

Despite medical and surgical advances, Fontan palliation of single ventricle birth defects remains problematic for a significant number of patients, leading to Fontan failure [1]. Patients with failing Fontan circulations have been implanted with a ventricular assist device (VAD) as a bridge to transplant [2,3]. Although systemic VAD support unloads the native ventricle, diminishing ventricular volume and external work, and augmenting the myocardial supply-demand ratio, its use in Fontan may not be ideal. In a Fontan circulation, preload is often insufficient and pathophysiology predominates on the right side of the circulation. While there have been reports of successful bridge to transplantation using VAD support in failing Fontan patients for brief periods of time, the results reported for post-cardiotomy bridge to transplantation have been poor [4]. Significantly, pediatric bridge to transplant support of failing Fontan patients with a VAD is usually not successful.

A concerted effort is currently underway to develop cavopulmonary assist devices (CPAD) to power the Fontan circulation by delivering a modest pressure boost (2–5 mmHg) at the level of the total cavopulmonary connection (TCPC) [5–9]. Support considerations include two microaxial pumps in the vena cava(e) [5] or a single percutaneous pump which can augment Fontan flow in all 4 axes of the TCPC without risk of venous pathway obstruction [6–8]. Once a safe and reliable device becomes available, it will be possible to provide high-volume, low-pressure flow augmentation similar to normal right ventricular hemodynamics to alleviate the sequelae of elevated systemic venous pressure and low cardiac output. In a univentricular Fontan circulation, CPAD support will simultaneously decrease systemic venous pressure and increase ventricular preload. It would restore physiologic status to one more closely resembling more stable 2-ventricle physiology, in essence enabling clinical management of the patient as a “biventricular Fontan” [1].

A key question to address prior to clinical application of mechanical circulatory support is how the single ventricle will respond to increased preload with CPAD support or reduced ventricular pressures and volumes with VAD support in the setting of longstanding systolic and/or diastolic dysfunction. A long-term follow up of Fontan survivors found preserved systolic function in 73% of subjects and diastolic dysfunction in 72% of patients [10,11]. Diastolic dysfunction is the predominant pathophysiologic feature in Fontan patients and is presumably secondary to: 1) prior staged repair in which a volume load was imposed on the ventricle; and 2) chronic preload deprivation and impaired ventricular filling. Thus, we hypothesize that a majority of patients with failing Fontan circulations will respond favorably to an increase in preload from cavopulmonary assist by increased cardiac output.

The circulatory response to VAD or CPAD support in the presence of systolic vs. diastolic dysfunction is undefined. Animal models of Fontan do not exist, making it a challenge to precisely define the circulatory response to cavopulmonary assist prior to clinical application. In this study, *in silico* and *in vitro* mock circulatory modeling were used to assess the circulatory response to VAD and cavopulmonary assist with respect to systolic and diastolic ventricular dysfunction. Based on the findings, an objective algorithm to guide clinical application of cavopulmonary assist is proposed.

Methods

Computer Simulation Model

A previously reported computer simulation model of the pediatric (~15–25 kg) single ventricle Fontan physiology was used in this study [7]. This Fontan model was developed from a biventricular computer simulation model that has been used in previous studies to develop and test physiologic control algorithms for mechanical circulatory support devices [12–15]. Briefly, the computer model subdivides the Fontan circulatory system into 2 heart valves and 9 blocks, which include common atrium, single ventricle, pulmonary and systemic circulations, vena cava, aorta, and coronary circulation. The volume of blood in each block is described by a differential equation as a function of volume (V), pressure (P), compliance (C), and resistance (R), which is an expression for the macrosopic material balance for the block given by:

$$\frac{dV_n}{dt} = F_n^{in} - F_n^{out}, \frac{dV_n}{dt} = \frac{V_{n-1}}{C_{n-1}R_{n-1}} - \frac{V_n}{C_n}\left(\frac{1}{R_{n-1}} + \frac{1}{R_n}\right) + \frac{V_{n+1}}{C_{n+1}R_n}.$$

where dV_n/dt is the rate of change of volume in block *n*, F^{in} is the blood flow rate into the block, and F^{out} is the blood flow rate out of the block.

The heart rate, resistances, and compliances of the Fontan model were modified to reproduce hemodynamic pressure and flow waveforms of the univentricular Fontan physiology of a 4 year old with (i) diastolic dysfunction with normal pulmonary resistance (NPR), (ii) diastolic dysfunction with elevated pulmonary resistance (Right/pulmonay side failure, RSF), (iii) systolic dysfunction due to diminished ventricular contractility and normal pulmonary resistance (left/systemic side failure, LSF), and (iv) combined systolic and diastolic dysfunction due to diminished ventricular contractility and elevated pulmonary resistance (left and right sided failure, LRSF) were developed based on literature [10,16–18] and clinical guidance. Specifically, RSF was simulated by increased pulmonary arterial and venous resistances, which resulted in reduced preload and venous return. LSF single ventricle Fontan circulation was modeled by adjusting the time-varying compliance curve of the single ventricle block to diminish ventricular contractility to simulate systolic dysfunction. LRSF single ventricle Fontan circulation was modeled via integration of increased pulmonary resistances from RSF and altered ventricular time-varying compliance curve from LSF.

Models of a VAD and/or CPAD were integrated into the computer simulation models of univentricular Fontan circulation. Simulations were conducted to predict acute hemodynamic responses including coronary flows, ventricular pressure-volume loops, ventricular external work, arterial pressures, and vascular pulsatility parameters for VAD and CPAD flow ranging from 0L/min (no support) to 3.25 L/min (full support). Ventricular, aortic, and cavopulmonary pressures, aortic, coronary, and cavopulmonary flows, and ventricular volume and external work were calculated.

In-vitro mock circulatory loop modelling

A mock circulation system consisting of a silicone ventricle, aorta, systemic and pulmonic resistances and compliances, and a cavopulmonary junction was used to simulate the univentricular Fontan circulation (Figure 1) [7]. The cavopulmonary junction is rigid with 11 mm diameter SVC and IVC and 9mm diameter pulmonary arteries that are connected to flexible silicone tubing. The clinically approved ventricular assist device (HeartWare HVAD, Miami Lakes, FL) inlet was attached to the single ventricle apex and the outlet cannula was attached to the proximal aorta. A CPAD (Viscous Impeller Pump, Indianapolis, IN) that is currently under development was placed in the cavopulmonary junction. Aortic, pulmonary arterial, and VAD flows were measured using Transonic Flow Probes (Transonic Systems, Ithica, NY). Aortic (proximal and distal), atrial, vena caval, and pulmonary arterial pressures were measured using single tipped pressure catheters (Millar Instruments, TX). The single ventricle pressure and volume were measured using a pressure-volume conductance catheter (Millar Instruments, TX). Ventricular driveline pressure, heart rate, systemic and pulmonary resistances and compliances were adjusted to reproduce hemodynamic waveforms of univentricular Fontan physiology of a 4-year old with (i) diastolic dysfunction with normal pulmonary resistance (NPR), (ii) diastolic dysfunction with elevated pulmonary resistance (RSF), (iii) systolic dysfunction due to diminished ventricular contractility and normal pulmonary resistance (LSF), and (iv) combined systolic and diastolic dysfunction due to diminished ventricular contractility and elevated pulmonary resistance (LRSF). RSF was simulated by increasing pulmonary resistance, which resulted in diminished preload and venous return. LSF single ventricle Fontan circulation was modeled by reducing the driveline pressure of the pneumatic driver to diminish ventricular contractility to simulate systolic dysfunction. LRSF single ventricle Fontan circulation was modeled by increasing pulmonary resistance and reducing ventricular driveline pressure. Baseline hemodynamic pressure and flow data were collected for the univentricular Fontan circulation (no VIP or VAD support). Hemodynamic data were obtained for partial (1.7±0.2 L/min) and full (3.2±0.3 L/min) VIP and/or VAD support with the VIP/VAD rpm adjusted to match the desired flow rate.

Results

Fontan circulation with diastolic dysfunction (NPR)

The unsupported Fontan circulation, even with normal pulmonary resistance, had elevated cavopulmonary pressure and diminished preload, cardiac output, aortic systolic and diastolic pressures compared to normal biventricular circulation [12], indicating diastolic dysfunction. CPAD support increased cardiac output, ventricular end-diastolic pressures and volumes, and aortic systolic and diastolic pressures and volumes (Table 1,Figure 2A,3A). CPAD support restored cardiac output, ventricular end-diastolic pressures, and aortic systolic and diastolic pressures to normal biventricular circulation values. Significantly, the restoration of hemodynamic parameters of the Fontan circulation to near normal values was achieved with only modest shift of pressure head (~5 mmHg) in the cavopulmonary junction. VAD support diminished ventricular external work and increased cardiac output. Importantly, ventricular end diastolic pressure and volumes were significantly diminished beyond nominal values indicating an increased risk of suction (Figure 2A,3A). Combined CPAD and VAD support

augmented ventricular systolic pressures but end-diastolic pressures and volumes were not augmented significantly from full VAD support values.

Fontan circulation with diastolic dysfunction and elevated pulmonary resistance (RSF)

Elevated pulmonary resistance resulted in significantly increased cavopulmonary pressure and diminished preload, cardiac output, aortic systolic and diastolic pressures compared to Fontan circulation with normal pulmonary resistance, Figure 2 B. CPAD support augmented cardiac output, ventricular end-diastolic pressures and volumes, and aortic systolic and diastolic pressures and volumes (Table 1,Figure 2B,3B). VAD support diminished ventricular external work and increased cardiac output. However, VAD support resulted in negative ventricular end diastolic pressures, which is a strong indication of suction (Figure 2A,3A). Combined CPAD and VAD support augmented ventricular systolic pressures but end-diastolic pressures and volumes were not augmented significantly from complete VAD support values.

Fontan circulation with systolic dysfunction (LSF)

Systolic dysfunction resulted in significantly increased preload and ventricular end diastolic pressures, and diminished cardiac output, aortic systolic and diastolic pressures compared to Fontan circulation with normal pulmonary resistance, Figure 2 C. The cardiac outputs of Fontan circulations with RSF and LSF were similar. VAD support diminished ventricular end diastolic pressure and preload to nominal values while augmenting cardiac output (Table 1,Figure 2C,3C). CPAD support augmented ventricular external work and increased ventricular end-diastolic pressures and volumes beyond normal values (Figure 2C,3C). Combined CPAD and VAD support diminished ventricular end-diastolic pressures and volumes compared to CPAD support values. However, the ventricular end diastolic pressures were still higher than the normal range.

Fontan circulation with systolic and diastolic dysfunction (LRSF)

Fontan circulation with systolic and diastolic dysfunction results in significantly diminished cardiac outputs compared to NPR, LSF and RSF, Table 1. VAD support caused ventricular suction while CPAD support increased ventricular end diastolic pressure above nominal values. Combined CPAD and VAD support resulted in diminished ventricular work, pressures and volumes. Importantly, combined CPAD and VAD support resulted in ventricular end diastolic pressures in the normal range.

DISCUSSION

Over the past 30 years, an increasing cohort of survivors of Fontan palliation of functional single ventricle is emerging with many of these patients expected to eventually present with clinical Fontan failure. The Fontan patient cohort has several subgroups with different etiologies and underlying causes for Fontan failure. Fontan failure is typically not the same as systemic ventricular failure with systolic dysfunction. A long-term follow up of Fontan survivors found preserved systolic function in 73% of subjects and diastolic dysfunction in 72% of patients [10,11]. The clinical manifestations of Fontan failure may be more representative of decompensated systemic venous sequelae of Fontan physiology rather than

that of primary ventricular failure. Stated more clearly, primary myogenic failure is not the underlying precipitating problem.

Currently, the therapeutic options for failing Fontan patients are limited to medical therapy, surgical optimization of passive Fontan flow, and mechanical circulatory support therapy. Medical therapy is of modest value, and only represents secondary therapy. Diuretic therapy may reduce the sequelae of increased tissue water, but at the expense of circulating blood volume, which is essential to maintenance of cardiac output and circulatory homeostasis. Inotrope therapy may increase ventricular contractility, but in a ventricle with insufficient preload, the magnitude of this benefit may be suboptimal. Phosphodiesterase inhibitors may have benefit on long-term functional status in prospective randomized trials, although the magnitude of this benefit is not yet clear [19]. To overcome the limitations of medical therapy, surgical approaches to passively optimize TCPC to reduce power losses by perhaps 1–2 mmHg have been proposed [20]. In this region of the circulation, an incrementally small improvement in hemodynamic efficiency may have tangible benefits. This is supported clinically by patients who undergo Fontan conversion from an atriopulmonary to a TCPC type of construction. However, surgical modifications of the TCPC have yet to be applied clinically on a significant scale. A very late surgical option is transplantation, which has issues and concerns of its own, and does not currently represent a long-term ideal option. For the forthcoming patients expected to present with Fontan failure, the lack of a primary therapy is increasingly a subject of urgency and concern.

Mechanical systemic circulatory support with VAD has been described for patients with failing Fontan circulation [2,3]. Systemic VAD support of Fontan can provide a reliable source of systemic flow, but it does not address the right-sided circulatory deficiencies typically inherent in Fontan circulation– namely coexisting systemic venous pressure elevation and reduced preload. To address this issue directly, a right-sided circulatory support device specifically designed to address these problems would be required. At present, a low-pressure device ideally suited to provide the 5–10 mmHg pressure necessary for transpulmonary blood flow is not commercially available. Applying contemporary VAD or total artificial heart (TAH) technologies to the right-sided Fontan circulation requires the take down of the Fontan connection to ensure effective right-sided flow [21]. The TCPC is an open venous channel, and there is no means to accomplish inflow and outflow cannulation in an unaltered TCPC without recirculation and lack of forward flow. Certainly, takedown of a Fontan connection and placement of bi-VADs or TAH is not a trivial consideration. TAH implantation also requires the creation of a receptacle for systemic venous return complicating surgical complexity and risk. Further, the device (VAD or TAH) becomes the sole, obligate path for right-sided cardiac output. If the device fails for any reason, it is a potentially lethal problem. Current MCS technologies are optimally designed to provide systemic support, and cannot address the unique needs of Fontan cavopulmonary support. For these reasons, the existing options to apply commercially available VAD support in a failing Fontan circulation are extremely limited and are associated with poor outcomes [4]. Emerging concepts in cavopulmonary assist devices (CPAD), however, may change this treatment paradigm by specifically addressing the right-sided circulatory support needs in a Fontan circulation [5–9].

A key question to address prior to clinical application of mechanical circulatory support in Fontan patients is how the single ventricle will respond to increased preload with CPAD support or reduced ventricular pressures and volumes with VAD support in the setting of longstanding systolic and/or diastolic dysfunction. The results of this study demonstrate that cavopulmonary assist improves failing Fontan circulation in the setting of diastolic dysfunction but preserved systolic function by augmenting cardiac output and increasing the preload to the single ventricle. Systemic VAD support is contraindicated for patients with diastolic dysfunction as it will further diminish preload and ventricular end diastolic pressure to unacceptably low levels which may precipitate ventricular suction. In the presence of systolic dysfunction and elevated ventricular end diastolic pressures, systemic VAD support augments cardiac output and diminishes ventricular end diastolic pressures. Increased preload with cavopulmonary assist may worsen circulatory status by significantly increasing the ventricular preload, volume, and end diastolic pressure. Inotropic support may offset the increased preload with cavopulmonary support to a certain extent. In cases of mixed systolic and diastolic dysfunction, both systemic and cavopulmonary support may provide the best outcome. Based on these findings, a mechanical circulatory support treatment algorithm for Fontan patients is presented in Figure 4. This algorithm is based on traditionally measured parameters for Fontan patients including ventricular end diastolic pressure, cardiac index, and pulmonary resistance. Pivotal to the selection of right-sided vs left-sided support is the underlying contractile status of the single ventricle.

Limitations

Chronic animal models of univentricular Fontan circulation that accurately replicate Fontan hemodynamics do not exist, making it a challenge to test the circulatory response to ventricular or cavopulmonary assist prior to clinical application. Computer simulation and mock circulation of the Fontan circulation is representative of clinical observations from a purely hemodynamic viewpoint, and is not intended to replace the importance and significance of *in vivo* models. While incapable of replicating all expected clinical conditions and responses, in-silico and in-vitro modeling does provide a controlled environment to test the effects of VAD and CPAD support and potential failure modes, which is valuable in device development and is not possible *in vivo*. As examples, the models do not simulate diastolic dysfunction due to restrictive atrioventricular valve. Systemic support with atrial cannulation will more likely to provide successful support in this condition. Additionally, diastolic dysfunction due to changes in end-systolic pressure volume relationship or isovolumetric relaxation time were not simulated. The computer simulation model does not account for the 2 mmHg respiratory variation in systemic venous pressure. Ventricular contractility and heart rate were kept constant to reduce experimental variability. Physiologically, heart rate and the contractility will increase with increasing preload in accordance with the Frank-Starling mechanism. The mock circulation system has mechanical valves which may create large aortic valve pressure gradients and ringing during valve closure. The computer simulation model does not account for viscosity changes or inertial effects while the length of tubing in the mock circulation may cause added inertial effects. However, the inertial effects represent less than 2% of the total power. Inertance mismatch or small viscosity changes would not affect the results significantly, as demonstrated by the similarity in results between the computer simulation and mock

circulation models. Despite these limitations, this study enabled the development of treatment algorithm using mechanical circulatory support devices for Fontan failure. These findings may be useful to guide clinical decision-making strategies for mechanical assist in patients with failing Fontan circulations in the future as Fontan-specific mechanical circulatory support devices come into clinical use.

Acknowledgments

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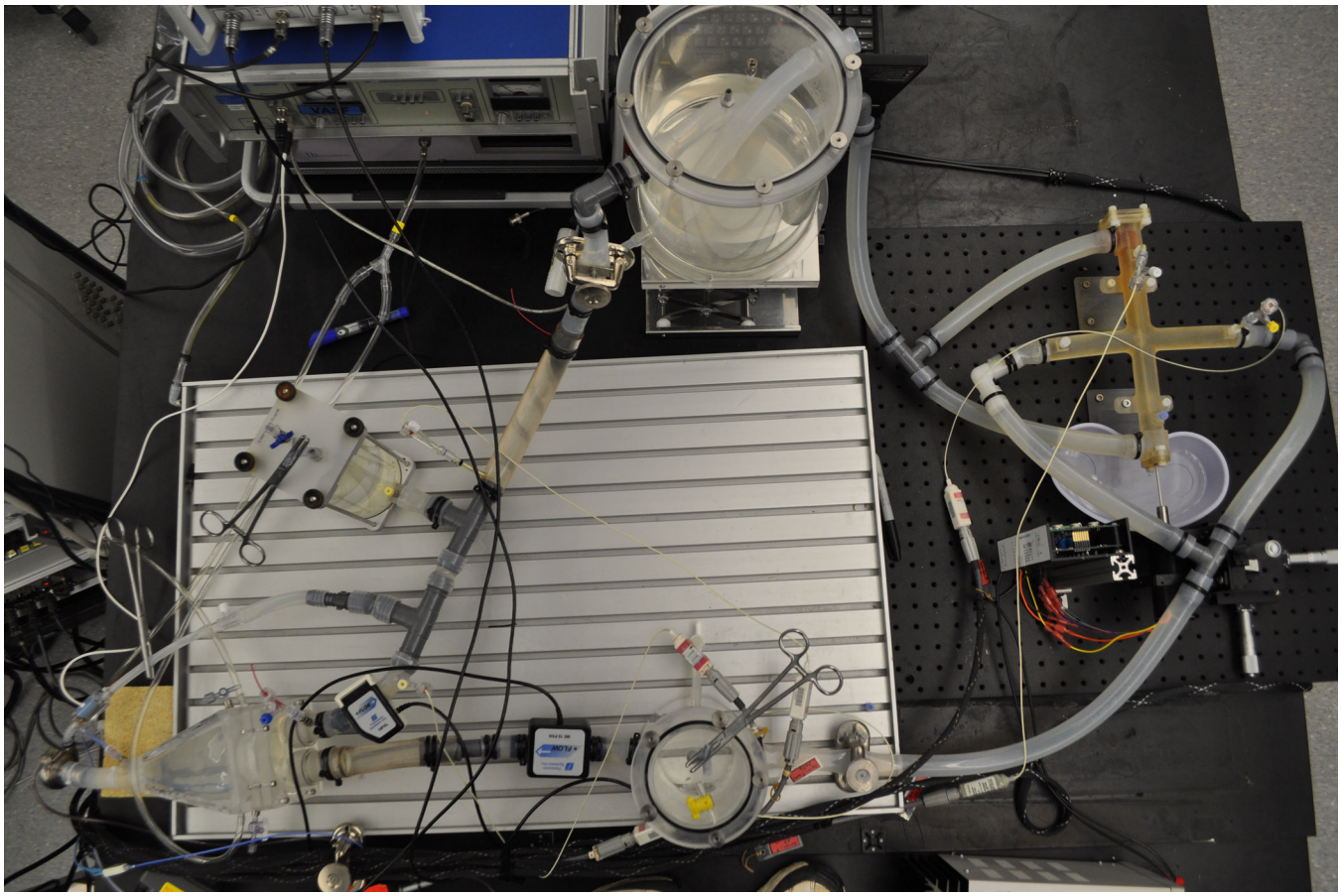
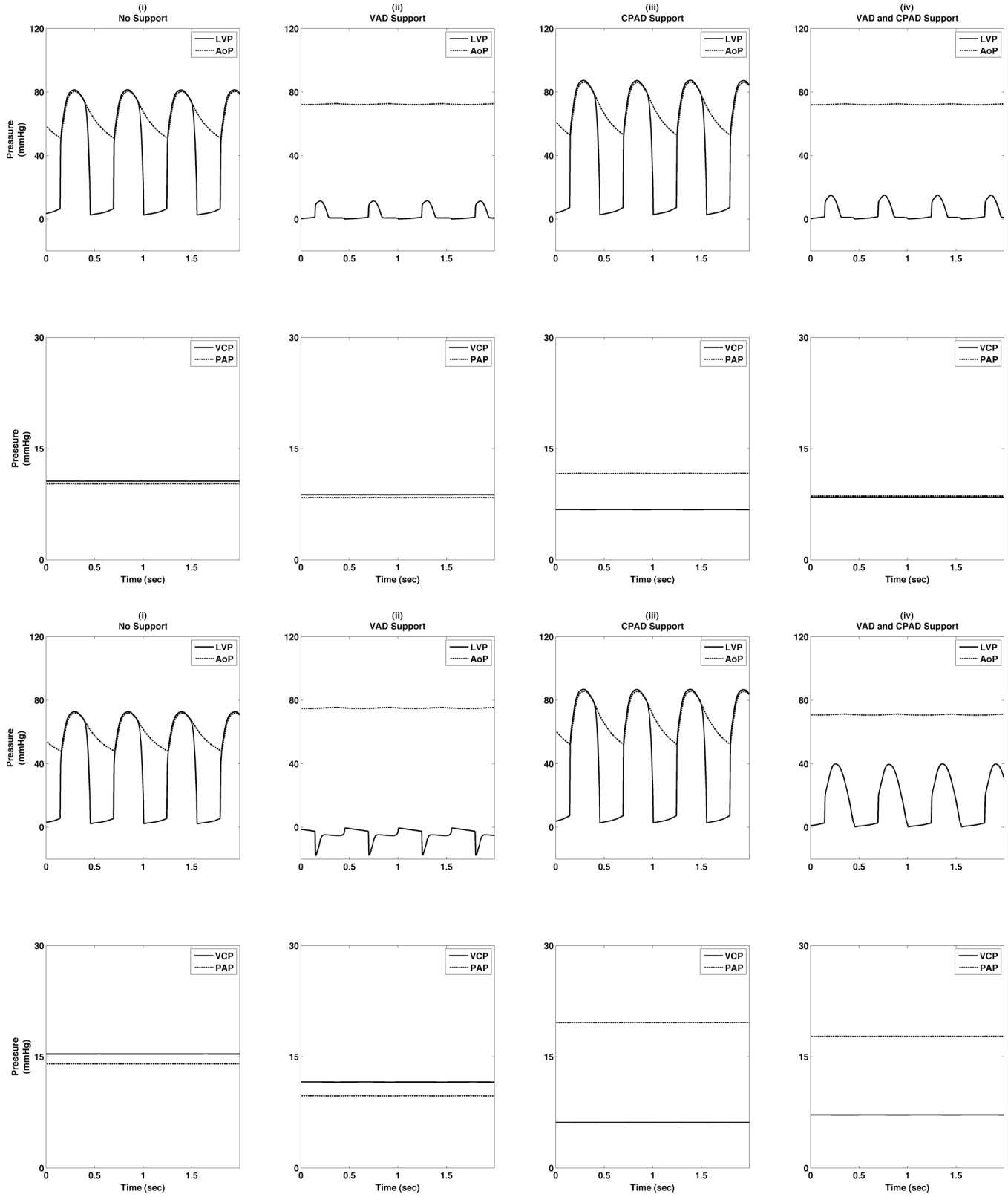
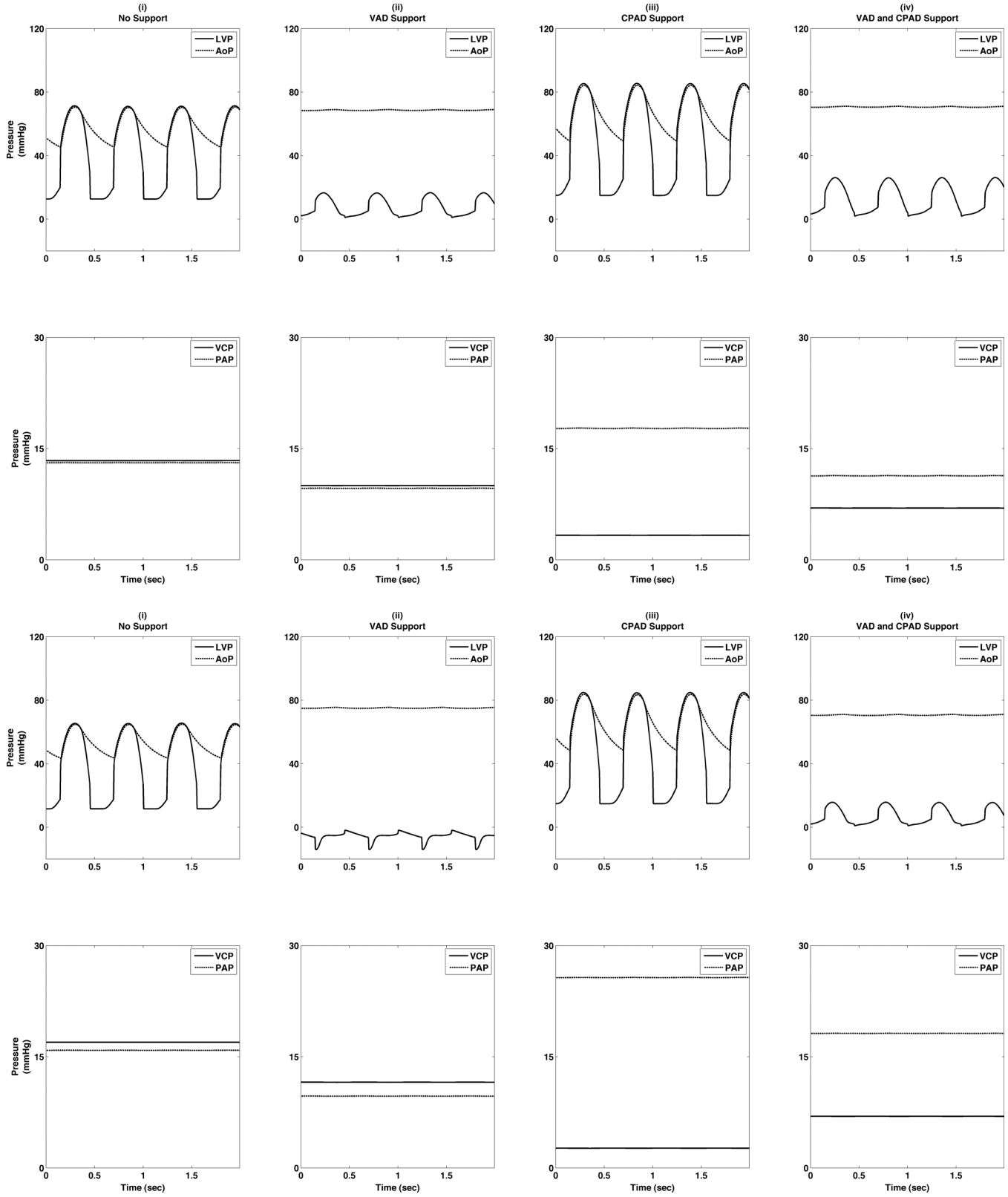


Figure 1.
Pediatric Fontan mock circulatory system with: (1) Single ventricle, (2) Aorta, (3) Arterial Compliance, (4) Systemic vascular resistance, (5) venous compliance, (6) Fontan junction with cavopulmonary assist device (VIP), (7) Pulmonary resistance, (8) Pulmonary compliance, and (9) ventricular assist device.





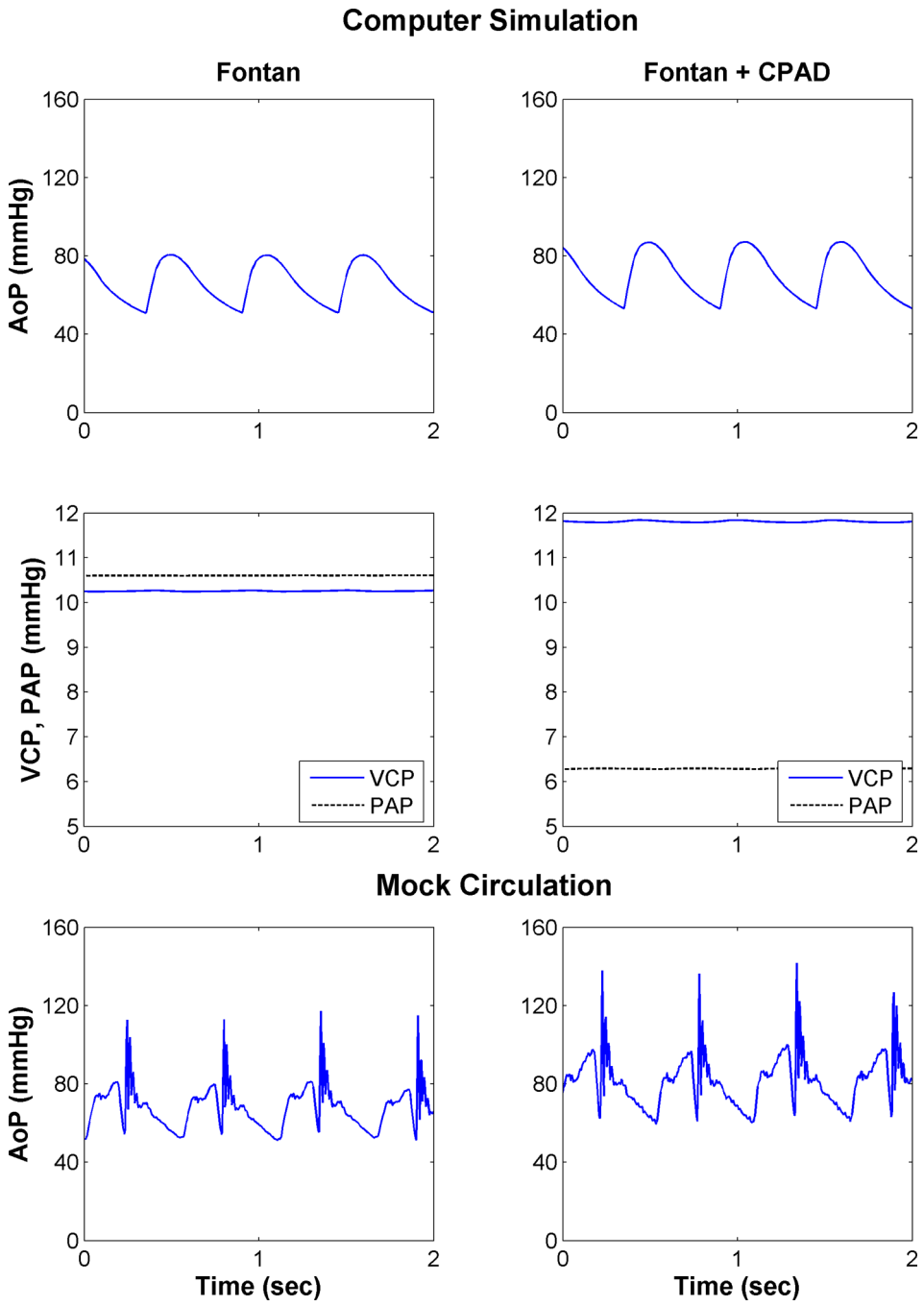
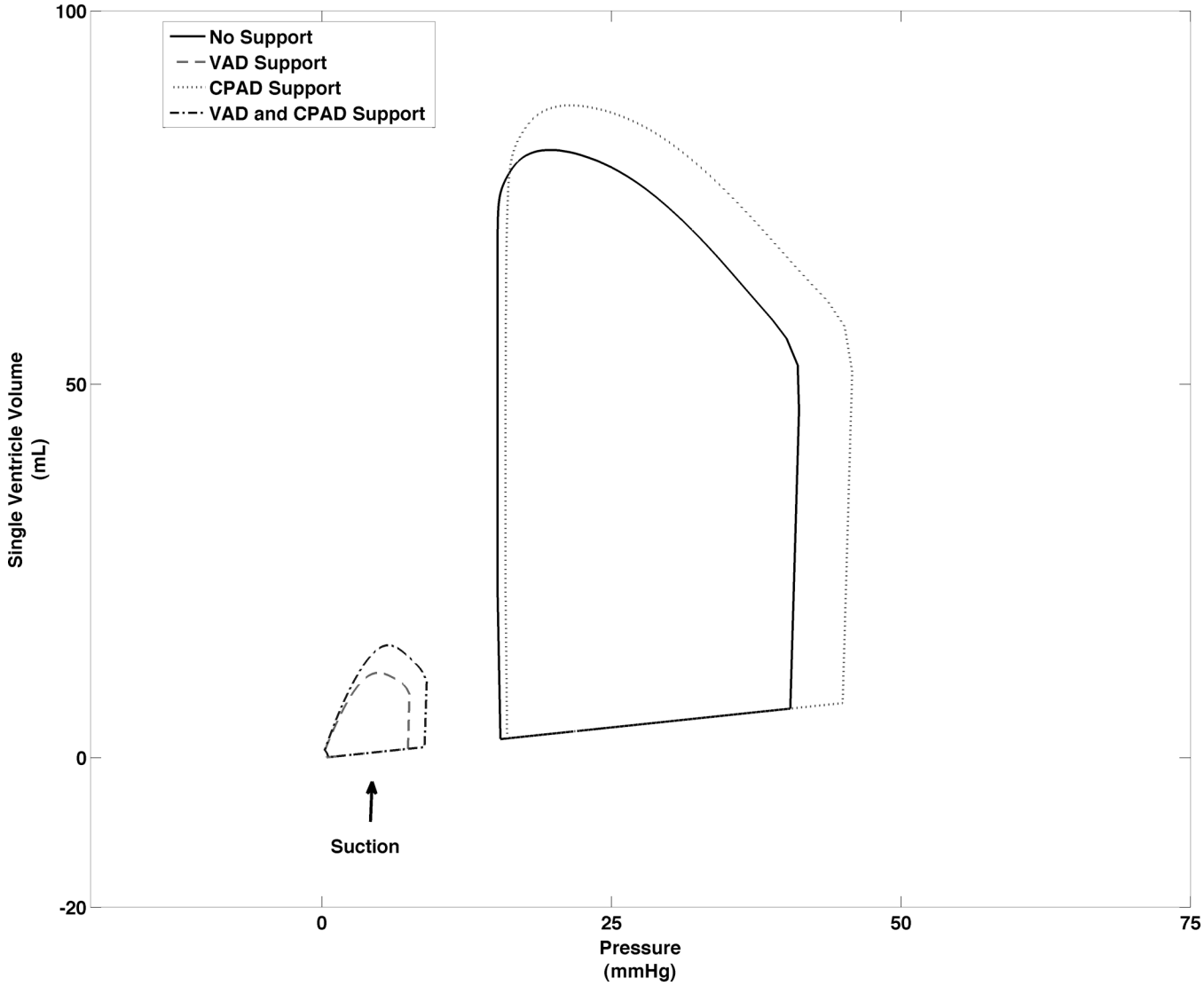
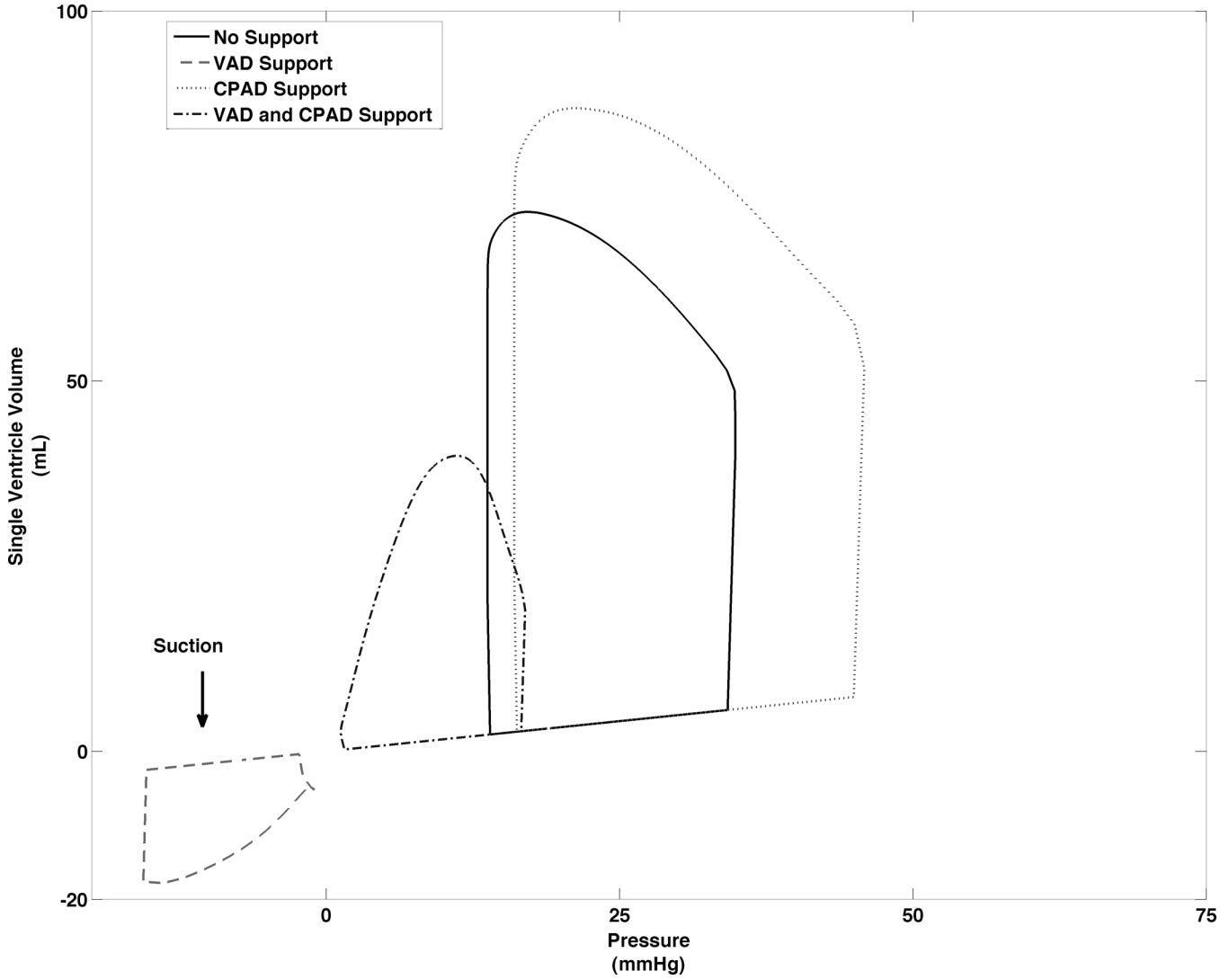


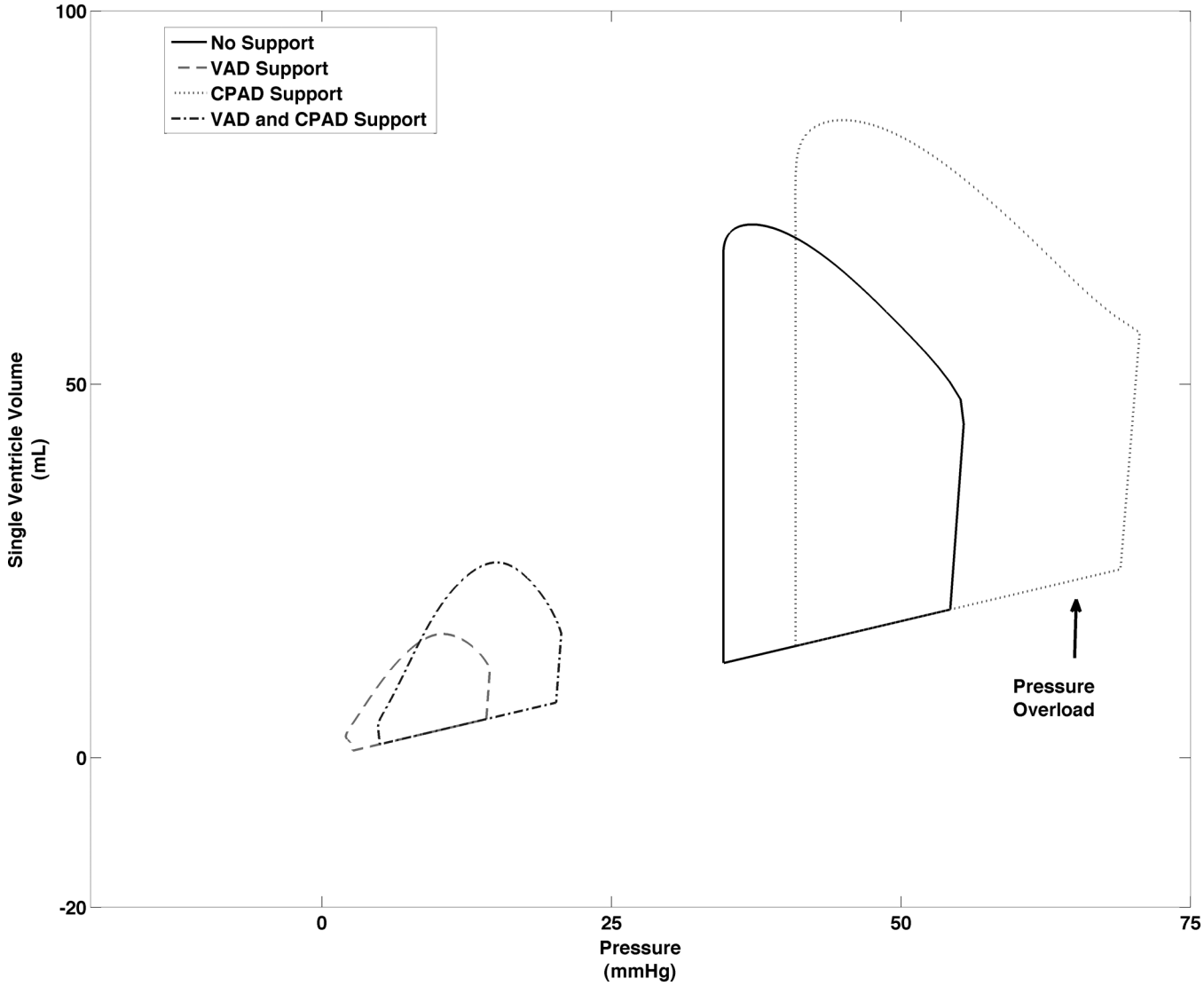
Figure 2.

Single ventricular pressure (SVP), aortic pressure (AoP), vena caval pressure (VCP), and pulmonary arterial vasculature pressure (PAP) during (i) no support, (ii) full VAD support, (iii) full CPAD support, and (iv) full VAD and CPAD in Fontan circulations with (A) normal pulmonary resistance (NPR), (B) diastolic dysfunction with elevated pulmonary resistance (Right/pulmonic side failure, RSF), (C) systolic dysfunction (left/systemic side failure, LSF), and (D) combined systolic and diastolic dysfunction (left and right side failure, LRSF). (E) Sample waveforms generated from computer simulation and mock

circulation demonstrate similar aortic pressures. The ringing in the aortic pressure waveforms are due to the tilting disc valve used in the mock circulation.







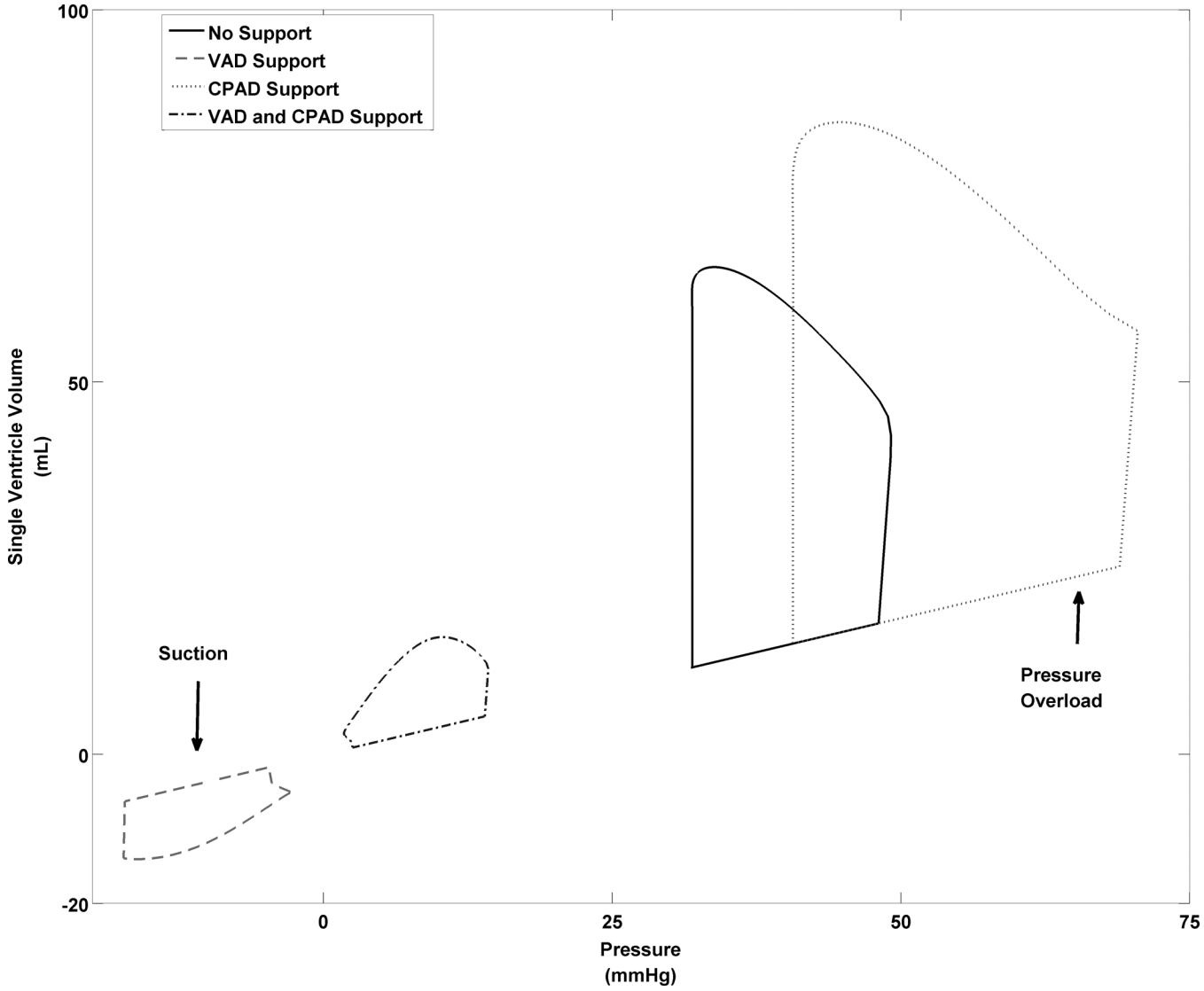


Figure 3.

Single ventricular pressure –volumes loops during no support, full VAD support, full CPAD support, and full VAD and CPAD support conditions in Fontan circulations with (A) normal pulmonary resistance (NPR), (B) diastolic dysfunction with elevated pulmonary resistance (Right/pulmonic side failure, RSF), (C) systolic dysfunction (left/systemic side failure, LSF), and (D) combined systolic and diastolic dysfunction (left and right side failure, LRSF).

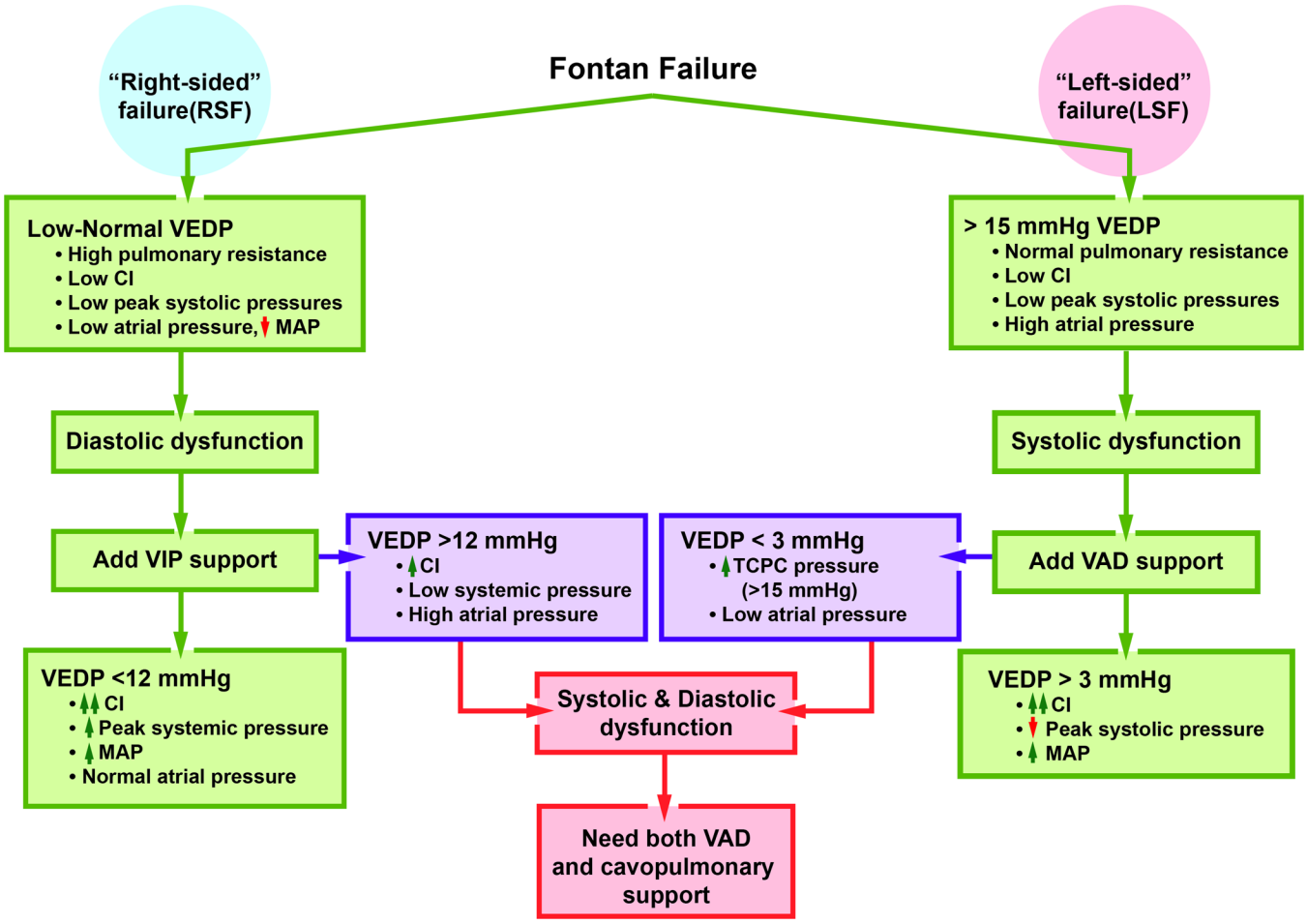


Figure 4.

Algorithm for mechanical circulatory support for patients with non-restrictive Fontan failure.

Table 1

Hemodynamic results for Fontan circulations during no support and various levels of CPAD and VAD support. The heart rate for all conditions was 110 beats per minute.

	VAD Flow (L/min)	CPAD Flow (L/min)	CO (L/min)	SV (mL)	Mean AoP (mmHg)	LVPed (mmHg)	Mean PAP (mmHg)	Mean VCP (mmHg)	CPPH (mmHg)
Diastolic dysfunction, Normal Pulmonary Resistance (NPR)	0.00	0.00	2.80	26.1	66.3	6.6	10.3	10.6	−0.4
	0.00	1.75	2.83	26.2	66.4	6.6	10.3	10.5	−0.1
	0.00	3.25	3.18	29.8	70.4	7.3	11.6	6.8	4.8
	1.75	0.00	2.89	19.4	67.4	5.1	10.0	10.3	−0.4
	3.25	0.00	3.25	7.4	72.4	1.2	8.4	8.8	−0.4
	3.25	3.25	3.25	16.2	71.2	3.9	10.4	7.6	2.8
Diastolic dysfunction, Elevated Pulmonary Resistance (RSF)	0.00	0.00	2.25	21.1	60.3	5.6	14.0	15.4	−1.3
	0.00	1.75	2.32	21.7	60.9	5.7	14.4	14.8	−0.4
	0.00	3.25	3.18	29.8	69.7	7.3	19.6	6.1	13.5
	1.75	0.00	2.34	14.3	61.3	4.0	13.8	15.2	−1.4
	3.25	0.00	3.25	14.5	75.1	−2.5	9.7	11.5	−1.9
	3.25	3.25	3.25	15.7	70.9	2.7	17.7	7.4	10.4
Systolic dysfunction (LSF)	0.00	0.00	2.24	20.8	57.7	19.8	13.1	13.4	−0.3
	0.00	1.75	2.26	20.9	57.9	19.9	13.2	13.2	−0.1
	0.00	3.25	3.27	29.7	66.8	25.3	17.7	3.3	14.4
	1.75	0.00	2.42	18.2	60.2	15.8	12.3	12.6	−0.3
	3.25	0.00	3.25	5.4	72.4	2.1	8.4	8.8	−0.4
	3.25	3.25	3.25	15.8	70.8	7.4	11.3	7.1	4.2
Systolic and diastolic dysfunction (LRSF)	0.00	0.00	1.87	17.3	53.8	17.6	15.9	17.0	−1.1
	0.00	1.75	1.91	17.9	54.3	17.9	16.3	16.4	−0.1
	0.00	3.25	3.27	29.8	66.3	25.2	25.7	2.7	23.0
	1.75	0.00	2.02	14.6	56.0	13.2	15.3	16.5	−1.2
	3.25	0.00	3.25	14.7	75.1	−6.3	9.7	11.6	−1.9
	3.25	3.25	3.25	12.5	70.7	5.1	18.2	7.2	11.0

VAD Flow: Ventricular assist device flow; CPAD flow: cavopulmonary assist device flow; CO: cardiac output; SV: stroke volume; AoP: aortic pressure; PAP: Pulmonary artery pressure; VCP: Vena cava pressure; CPPH: cavopulmonary pressure head;